

CAB19961

Product Information

Product SKU:	CAB19961	Gene ID:	1476	Size:	20uL, 100uL
Clone No:	-	Host Species:	Rabbit	Reactivity:	Human,Rat

Additional Information

Observed MW:	13kDa	Conjugate:	Unconjugated
Calculated MW:	11kDa	Isotype:	IgG

Immunogen Information

Background:	The cystatin superfamily encompasses proteins that contain multiple cystatin-like sequences. Some of the members are active cysteine protease inhibitors, while others have lost or perhaps never acquired this inhibitory activity. There are three inhibitory families in the superfamily, including the type 1 cystatins (stefins), type 2 cystatins and kininogens. This gene encodes a stefin that functions as an intracellular thiol protease inhibitor. The protein is able to form a dimer stabilized by noncovalent forces, inhibiting papain and cathepsins I, H and B. The protein is thought to play a role in protecting against the proteases leaking from lysosomes. Evidence indicates that mutations in this gene are responsible for the primary defects in patients with progressive myoclonic epilepsy (EPM1). One type of mutation responsible for EPM1 is the expansion in the promoter region of this gene of a CCCC GCCCGCG repeat from 2-3 copies to 30-78 copies.
Recommended Dilution:	WB, 1:500 - 1:2000 IF/ICC, 1:50 - 1:200
Synonyms:	PME; ULD; CST6; EPM1; STFB; CPI-B; EPM1A; TB
Purification Method:	Affinity purification
Immunogen:	Recombinant fusion protein containing a sequence corresponding to amino acids 1-98 of human CSTB (NP_000091.1).
Storage:	Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.01% thimerosal, 50% glycerol, pH 7.3.